

Sickle Cell Disease

A guide for people with sickle cell disease



**Ontario
Health**

Ontario Health is committed to improving the quality of health care in the province in partnership with patients, health care professionals, and other organizations.

To do that, Ontario Health develops quality standards. These are documents that outline what high-quality care looks like for conditions or processes where there are large differences in how care is delivered, or where there are gaps between the care provided in Ontario and the care patients should receive. These quality standards set out important steps to improve care. They are based on current evidence and input from an expert committee that includes patients, health care professionals, and researchers.

This patient guide accompanies the quality standard on [sickle cell disease](#). It outlines the top eight areas where providers can take steps to improve care for children, young people, and adults with sickle cell disease. The patient guide also includes suggestions on what to discuss with your health care providers, as well as links to helpful resources.

If you're a caregiver:

If you're helping to care for someone with sickle cell disease, this whole guide is for you, too—you may need this information as much as they do. We also recognize the unique concerns you might have as a caregiver as you support someone with a chronic condition. If you're caring for someone with sickle cell disease, their needs will change over time based on how they're feeling and the severity of their condition.

Below is a summary of the top eight areas to improve care for people with sickle cell disease.



Quality Statement 1: Racism and Anti-Black Racism

What the standard says

People with sickle cell disease (and their families and caregivers) experience care from health care providers within a health care system that is free from racism and anti-Black racism, discrimination, and stigma. Health care providers promote a culture that is compassionate, trauma informed, and respectful of people's racial/ethnic and cultural backgrounds. They build trust with people with sickle cell disease (and their families and caregivers), work to remove barriers to accessing care, and provide care equitably.

What this means for you

- Your health care providers should always treat you with respect, dignity, and compassion.
- You should be given the opportunity to be as healthy as possible. This means that you should be able to get high-quality health care when you need it, no matter where you seek care (for example, at your doctor's office or at the hospital).



Quality Statement 2: Comprehensive Health Assessment and Care Plan

What the standard says

People with sickle cell disease have a comprehensive health assessment at least annually from an interprofessional care team at a dedicated sickle cell disease centre to develop an individualized, person-centred care plan. The assessment and care plan are documented and shared with the person's circle of care.

What this means for you

- You should be offered a complete health assessment at least once a year. This involves checking your physical health, your mental health, and your overall well-being.
- Your health assessment should be done at a health care centre that specializes in treating people with sickle cell disease.
- Your health care provider should use this health assessment to make a plan for your care. They should share this plan with all your other health care providers so that everyone has the information they need to give you the best care for your sickle cell disease.

At your yearly complete health assessment, your health care provider should:

- Ask you about any pain you might be experiencing
- Provide education about the complications of sickle cell disease and tell you what early signs to watch out for
- Tell you about the treatment options available to you, including their benefits and risks
- Offer you information about other services or supports that might help you
- Tell you about any research studies you might be eligible for and help you find out how to participate if you would like to



Quality Statement 3: Vaso-occlusive Acute Pain Episodes

What the standard says

People with sickle cell disease who present to an emergency department or hospital with a vaso-occlusive acute pain episode receive a timely pain assessment and clinical assessment. Their treatment begins within 30 minutes of triage or 60 minutes of presentation. Before discharge, they are involved in the development of a plan for continuing to manage their acute pain episode at home. This plan includes symptom management strategies and information on how to access follow-up care and support from health care providers, as needed.

What this means for you

If you have to go to the emergency department or hospital for treatment for pain caused by sickle cell disease, a health care professional should see you and start treatment quickly:

- Within 30 minutes of being assessed by a nurse *or*
- Within 60 minutes of arriving at the hospital

Before you leave the hospital, a health care provider should work with you to make a plan to continue managing your pain at home. This plan is called a **discharge information sheet**.

Your health care provider should give you a copy of your discharge information sheet. They should provide it in a language you understand or offer translation or interpretation services if you need them.

Your health care provider should also give your discharge information sheet to a health care professional you see regularly for your sickle cell disease, like your family doctor, your nurse practitioner, or a doctor who specializes in treating people with sickle cell disease.

Your discharge information sheet should include information on:

- The treatment you received at the hospital, such as hydroxyurea therapy or pain medication
- How to continue managing your pain at home
- How to get more medication if you need it
- How to manage any medication side effects
- How and when to get a follow-up appointment if you need one
- What to do and who to contact if your pain becomes unmanageable

What can cause an acute pain episode?

- Dehydration (not drinking enough water)
- Being cold
- Being sick
- Having a fever
- Having surgery
- Stress in your life

Some people with sickle cell disease might worry that physical activity can cause an acute pain episode. It is important to exercise, so your health care provider should talk with you about this and help you understand how you can exercise safely.

Medications called **opioids** are often used to treat acute pain episodes. If you would like to learn more about how opioids are used to treat acute pain, please see Ontario Health's [*Opioid Prescribing for Acute Pain*](#) quality standard.



To learn more about high-quality care for people leaving the hospital, please see Ontario Health's [*Transitions Between Hospital and Home*](#) quality standard.



Quality Statement 4: Life-Threatening Acute Complications

What the standard says

People who present to an emergency department or hospital with a potentially life-threatening acute complication of sickle cell disease have their condition and its severity identified through a prompt clinical assessment. Their condition is managed appropriately with an individualized treatment and monitoring plan.

What this means for you

- Call 9-1-1 or go to your nearest hospital if you suddenly feel weak or if you have a fever, chest pain, a cough, trouble talking, or trouble breathing—even if you are not in pain.
- At the hospital, a doctor should see you quickly to check if you might be having a serious complication of sickle cell disease.
- Your doctor should talk with you about your treatment options and let you know the risks and benefits of each one. They should also ask if you have had any bad reactions to treatments or medications in the past.
- Your doctor should work with you to choose your treatment.

It is important to know your **baseline hemoglobin value**.

Hemoglobin is a protein in your red blood cells that helps them carry oxygen from your lungs to other parts of your body.



Your baseline hemoglobin value is the amount of hemoglobin you usually have in your blood. A health care professional you see regularly for your sickle cell disease should tell you what your baseline hemoglobin value is.

This information can help your health care professional at the hospital decide which treatment would be best for you.



Quality Statement 5: Chronic Complications

What the standard says

People with sickle cell disease are monitored by their local health care team for signs and symptoms of chronic complications of sickle cell disease. People with chronic complications are promptly referred to a dedicated sickle cell disease or other specialized centre for consultation and/or assessment and management of their condition, as needed.

What this means for you

- Chronic complications of sickle cell disease can have a serious effect on your health.
- Your health care provider should tell you what to watch out for so you can get treatment quickly if you need it.
- If you are experiencing a chronic complication, your health care provider should talk with health care professionals at a health centre that specializes in treating people with sickle cell disease to make sure you get the best treatment possible. They may also arrange for you to have an appointment at this centre.

Signs and symptoms of chronic complications to watch out for:

- Chronic pain (pain you have all the time)
- Leg ulcers (sores on your legs that take a long time to heal)
- Trouble with your kidneys, lungs, or eyes
- Anxiety (worrying a lot)
- Depression (feeling sad most of the time)
- Feeling lonely or helpless
- Not being able to sleep

To learn more about treatment for chronic pain, anxiety, and depression, please see Ontario Health's [Chronic Pain](#), [Anxiety Disorders](#), and [Major Depression](#) quality standards.





Quality Statement 6: Referral to Health Care Professionals With Expertise in Chronic Pain

What the standard says

People whose quality of life is significantly impacted by chronic pain caused by sickle cell disease are referred to individual health care professionals or a chronic pain centre with expertise in chronic pain related to sickle cell disease and the ability to offer pharmacological and nonpharmacological interventions.

What this means for you

If chronic pain is having a negative effect on your life, your health care professional should arrange for you to have an appointment with a doctor or at a health centre that specializes in treating chronic pain caused by sickle cell disease.

If you're a caregiver:

You might find it difficult sometimes to help the person you are caring for manage their chronic pain (pain they feel all the time). It is important that you and the person you are caring for work together with their health care professionals to decide on the best plan for managing their pain.

It is also important that you have all the information you need to help the person you are caring for. If you have questions, it can be helpful to write them down and bring them with you to health care appointments so you do not forget them.

To learn more about treatment for chronic pain, please see Ontario Health's [Chronic Pain](#) quality standard.





Quality Statement 7: Psychosocial Assessment, Information, and Support

What the standard says

People with sickle cell disease (and their families and caregivers, where appropriate) have regular psychosocial assessments to identify any psychosocial needs or barriers to accessing care. Those with unmet psychosocial needs are offered information and support to address these needs.

What this means for you

Living with sickle cell disease can be hard. So at least once a year, your health care provider should ask you about your mental health and your overall well-being. This includes asking if:

- You feel sad or worried a lot of the time
- You have trouble sleeping
- You feel lonely
- You are worried about school or work

Once your health care provider knows how you're feeling, they can let you know about services and supports that might help you if you would like that.

If you're a caregiver:

You might also have difficulties from time to time. This means that a member of the health care team of the person with sickle cell disease should also ask you about your mental health and well-being at least once a year. They should also offer you information and support that might help you if you would like that.



Quality Statement 8: Transition From Youth to Adult Health Care Services

What the standard says

Young people with sickle cell disease have a designated health care provider for the transition from youth to adult health care services. This provider works with the young person (and their parents and caregivers, where appropriate) to coordinate their care and provide support throughout the transition process. The provider continues to provide support until the young person (and their parents and caregivers, where appropriate) confirms that the transition is complete.

What this means for you

- Your health care providers should involve you in choosing a single provider to be the **designated health care provider** for your transition from youth to adult health care services. This should be someone you know and trust.
- This person should work with you to coordinate your move to adult services.
- They should help arrange appointments and provide support until you feel your transition is complete.

Your **designated health care provider** is one person from among your health care providers who agrees to coordinate your move to adult services. You can help decide who this provider is. If this move takes a long while, over time you might have more than one designated health care provider.



To learn more about transitions from youth to adult health care services, please see Ontario Health's [Transitions From Youth to Adult Health Care Services](#) quality standard.



Suggestions on what to ask your health care providers to help you receive high-quality care

- How do I know if I am having an acute pain crisis or other complication of sickle cell disease, and when do I need to go to the hospital?
- What is my baseline hemoglobin value?
- What can I do to prevent or manage acute and chronic pain at home?
- What treatment options are available to me, and what are their benefits and risks?
- What types of community and social supports are available to me and to my family and caregivers? (Examples of social supports include counselling, peer support, and financial benefits.)
- What mental health services are available to me and to my family and caregivers?
- What services and supports are available to help me with things like education, employment, finances, or housing?
- What can I do or say to make sure I get the help I need when I'm at the emergency department or at a doctor's office?
- Who can I contact if my family, caregivers, or I am being mistreated or if we are not receiving the care we need?

Suggestions on what to share with your health care providers to help you receive high-quality care

- The type of information and support you want from your health care team
- Your baseline hemoglobin value
- How you are feeling overall and what it's like for you and your caregivers to manage your sickle cell disease
- If you or your caregivers are feeling overwhelmed and need extra support
- If you are experiencing any new pain, if pain has come back, or if you are experiencing any other complications of sickle cell disease
- If you have a fever or other symptoms that could mean you have an infection
- The medications you are taking
- Any medications or treatments that don't work for you
- Any care plans you have (take these with you when you visit the emergency department or see a new health care provider)
- If you want to make changes to your medication, treatment, or care plan
- If you are planning to have children
- If you have any travel plans (if you are flying or if you are travelling outside the province, it is important to make sure that you will have the medication you need and that you know what to do if you need health care while you are away)



Learn more

The [American Society of Hematology](#) provides information on hydroxyurea therapy.

[ASE Community Foundation for Black Canadians with Disability](#) aims to bridge the knowledge and service gap in health care for Black Canadians with critical research and partnerships centring on disability justice and the unique experiences of Black Canadians with disabilities.

The [Centers for Disease Control and Prevention](#) provide information and resources, including fact sheets, infographics, videos, and podcasts.

The [Patient Ombudsman](#) provides free and confidential services to people who have already tried to have a complaint about health care services addressed by a public hospital, a long-term care home, or Home and Community Care Support Services and are not satisfied with the outcome.

The [Red Blood Cell Disorders Hub](#) is an online space for people to connect about sickle cell disease and other red blood cell disorders in Ontario.

The [Sickle Cell Association of Ontario](#) provides support and resources such as information on [sickle cell disease](#) and educational events.

The [Sickle Cell Awareness Group of Ontario](#) is a provincial organization that provides information on programs, community services, supports, and educational resources. Parents and caregivers can also find helpful resources, including information on [pain management](#) and [physical activity](#).

The Hospital for Sick Children has created [Sickle Cell Disease: A Practical Guide for Parents](#) and [Sickle Cell Disease: A Practical Guide for Teachers](#), which provide an easy-to-understand overview of sickle cell disease for parents and teachers.

Need more information?

If you have any questions or feedback about this guide, please contact us at QualityStandards@OntarioHealth.ca or 1-877-280-8538 (TTY: 1-800-855-0511).